sighted child (Adelson and Fraiberg, 1976) but is not motivated to explore and practice his motor skills and, of course, he will not practice any of the visual-motor skills in learning object permanence, object manipulation and spatial awareness due to his lack of vision.

Body image plays a significant role in the development of both motor and perceptual skills. It is through movement, firstly reflexive and then spontaneous active movement that the infant learns about his body, his control over it and its relationship to the environment. External influences, such as the way parents hold, wrap, hug and cuddle, provide sensations that help establish body boundaries and a separation of the body from objects.

Visually impaired children have difficulty in acquiring spatial orientation concept necessary for proficient locomotion and independent mobility. These children do not generally have retarded motor development but have lags in development and are not motivated to move freely, hence their motor skills are not as good or coordinated as their sighted peers, at school entry age of five years. There are many factors that may contribute to these facts, such as overprotection, restricted activity, lack of adequate stimulation and practice. Early stimulation needs to be analysed and goal oriented so as not to over-stimulate the young child who may be overcome by too much information and opt out to withdraw into himself. Adelson & Fraiberg (1976) found that blind infants develop similar patterns of neuromuscular maturation but that visually impaired infants exhibited delays in self-initiated mobility and locomotion. Griffin (1981) supports this view and stated that motivation associated with locomotion was slower in developing because of lack of visual input for assurance in space and walking cues.

On examining the movements and patterns of locomotion in 5-6 year old visually impaired children I have found that they demonstrated trunk instability, poor posture, incoordinated movements, small steps, abnormal arm postures and 'blindisms'. Many of these children are unable to hop, skip or jump, and on closer examination are not proficient in rolling, crawling, or sound location and fine motor skills.

This paper proposes that there is a need for parent education and advice from the physiotherapist on handling the visually impaired infant and that motor development programmes for visually impaired pre-school children are essential for optimal development. By starting at an early age the physiotherapist aims at developing the necessary readiness skills that underlie total body awareness and space through movement. Thus the visually impaired child at school entry age would have the basic skills equal to his sighted peers and be able to concentrate on maximising his potential in school.

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CT SCAN AND DOPPLER FLOW CHANGES WITH VENTRICULO-PERITONEAL SHUNTING

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Ventricular size on CT scan is widely used in asessing shunt function in hydrocephalus. The effect of raised intracranial pressure on cerebral blood flow may provide a more useful monitor of shunt function.

Transcranial doppler measures blood flow velocity in the middle cerebral artery using a range gated ultrasound beam directed from the temporal region. Measurements of systolic (S), diastolic (D) and mean (M) flow velocity are obtained. The pulsatile index (PI), which is a measure of vascular impedance, is derived from these values ($PI = {}^{S-D}/_{M}$).

The PI has been measured pre and post operatively in nine patients (age range 3 days - 4 years) requiring shunts for hydrocephalus. CT scans were performed on the same day. Indices of ventricular dilatation were obtained for the frontal and occipital horns of the lateral ventricles from coronal sections at the level of third ventricle. The intra ventricular pressure was determined by manometry at insertion of the shunt.

There was clinical evidence of shunt patency in all cases and in eight of the nine a reduction of pulsatility index. This implies diminished vascular impedance with shunting. There was a positive correlation ($r=0,68$, $P<0.05$) between change in pulsatility index and intraventricular pressure. No correlation existed between CT scan changes and intraventricular pressure or between CT scan and changes in pulsatility index.

The study demonstrates a greater correlation between pressure and pulsatility index rather than CT scan changes. This suggests that Doppler flow assessment may more accurately reflect shunt function.

CLINICAL VALUE OF VISUAL EVOKED RESPONSES IN CHILDREN

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Between January 1985 and December 1987 a total of 296 VER studies were performed on infants and children in our Depatrment. Cz and Fz (10/20 system) were used as reference and neutral electrodes with Oz as active. Binocular flash stimulation was performed in under five year olds and pattern reversal for older patients. Most patients also had conventional EEG studies and an ERG using a noncorneal electrode on the bridge of the nose

Results	FLASH VER CONTROL VALUES	
	N2	P2
0-4 mth.	88.8 (sd 25.1)	142.6 (sd) 29.4)
4 mth - 1 yr	64.3 (sd 3.1)	97.0 (sd 3.7)
1 yr - 4 yrs	70.0 (sd 9.9)	110.7 (sd 9.8)
4 yrs - 13 yrs	67.3 (sd 9.7)	108 (sd 11.8)
	PATTERN VER CONTROL VALUES	
	P 100	
5 yrs +	99.9 (sd 4.0)	

Abnormal VER studies were found in a variety of neurological disorders particularly progressive neurological diseases including Leighs Disease, Ceroidlipofuscinosis, Canavan's Disease and other Leucodystrophies. Our preliminmary data would indicate that the VER coupled with ERG and EEG was useful in the investigation of a child with a suspected progressive neurological disorder. Our experience also confirms that serial VER's are a useful predictor of visual outcome in infants with clinically suspected visual failure following acute /infectious and/or metabolic encephalopathy.

KAWASAKI DISEASE – THE JAPANESE PERIL

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Although first describe over twenty years ago, there is as yet no test available for the diagnosis of Kawasaki disease. Clinical criteria for the diagnosis of this condition have been laid down by the Japanese Kawasaki Disease Committee (Japanese Kawasaki Disease Research Committee: Diagnostic Guidelines of Kawasaki Disease. 4th ed. 1984, Tokyo, Japan).

Two children whose illness initially fulfilled the clinical criteria for the diagnosis of Kawsaki disease were subsequently found to have acute bacterial endocarditis on the one hand and a disseminated non-Hodgkin's lymphoma on the other. We describe their clinical course and emphasise the importance of other illnesses which may have a similar presentation as well as strict adherence to the criteria for Kawasaki disease.

LONG-TERM TREATMENT OF HYPOGLYCAEMIA DUE TO NESIDIOBLASTODSIS USING SOMATOSTSATIN ANALOGUES SMS 201-995

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Hyperinsulinism due to nesidioblastosis is the commonest cause of persistent hypoglycaemia in infants over one week of age. To avoid cerebral damage, the treatment of choice is early sub-total

pancreatectomy; if unsuccessful, infants require total pancreatectomy.

Aim : To use subcutaneoius somatostatin analogues (SMS 201-995) in the long-term treatment of recurrent hypoglycaemia in an infant with nesidioblastosis after sub-total pancreatectomy.

Case : An infant girl suffered recurrent hypoglycaemia despite dextrose infusion, diaxode and prednisolone. Inappropriately high insulin concentrations during hypoglycaemia and hypoketonaemia confirmed hyperinsulinism. At six weeks of age, subtotal pancreatectomy was performed; the histological diagnosis was nesidioblastosis. Hypoglycaemia recurred post-operatively despite continued dextrose infusion.

Method and Results : Subcutaneous somatostatin analogue was given in varying doses as a therapeutic trial. The effects on hourly glucose, beta-hydroxybutyrate and insulin during the first day of treatment were compared to the relevant concentrations over the preceding day. Two hours after the first dose, blood glucose increased from 0.2 mmol/l to 10 mmol/l, beta-hydroxybutyrate increased from 0.04 mmol/l to 0.22 mmol/l and serum insulin fell from 1.7 mU/l to 0.2 mU/l. With continued treatment of subcutaneous injections of SMA 201-995 less frequent episodes of hypoglycaemia occurred. At eighteen months of age, after sixteen months continuous treatment, both growth velocity and developmental screening were normal.

Conclusion : Subcutaneous somatostatin analogue is a safe and effective means of helping control severe hypoglycaemia, not only in the short term, but also as long-term therapy.

TRANSIENT HYPERPHOSPHATASEMIA OF INFANCY AND EARLY CHILDHOOD

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Transient Hyperphosphatasemia is a benign biochemical abnormality which occurs in children up to five years of age. It is characterised by a marked elevation in serum alkaline phosphatase, of less than three months duration, without evidence of hepatic or skeletal disease. There have been no consistent clinical findings in the 114 children reported with this condition to date.

Transient hyperphosphatasemia was a chance finding in three children admitted to our hospital with respiratory infections. Age range was from two to five years.

Electrophoresis of serum from these patients demonstrated the normal alkaline Isoenzyme bands and the two additional bands characteristic of serum from children with transient hyperphosphatasemia. These additional bands of a heat-labelled cathodal isoenzyme which probably derives from bone and a second isoenzyme with similar appearance and heat stability to the hepatic isoenzyme, but, demonstrating atypical anodal migration. It has been suggeated that the atypically-migrating anodal band represents an excessively sialylated isoenzyme of liver origin and that the excessive sialylation represents a response to a virus. The virus may alter the structure of the hepatic isoenzyme of alkaline phosphatase contributing to the impaired clearance from blood.

Transient hyperphosphatasemia is usually an incidental finding and does not require treatment or further investigation. Follow-up is desirable as long-term sequelae, while thought to be negligible, are unknown.

BACTERIAL TRACHEITIS

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Bacterial Tracheitis is an uncommon condition characterised by severe stridor, extreme toxicity and the presence of thick muco-purulent material in the subglottis and trachea. The lining mucosa is grossly oedematous and there may be accompanying submucosal haemorrhages.

In this study, 4 children between the ages of 5 and 6 years, treated at the Children's Hospital, Temple Street for bacterial tracheitis over a one year period are reviewed.

All 4 patients presented with a 48-72 hour history of mild hoarseness, brassy cough, followed by rapidly progressive stridor, fever and toxicity. One patient had positional preference. All patients required direct laryngoscopy.

At endoscopy the epiglottis and supraglottis was normal in all cases. All had marked oedema of the subglottis and trachea, with severe submucosal haemorrhages in 2 patients.

All patients had thick muco-purulent material in the subglottis and upper trachea which was cleared by suction clearance at the time of bronchoscopy. Three patients required endotracheal intubation. All patients were admitted to I.C.U., though none required ventilation. Intubated patients had repeated suction clearance of tracheal secretions. All were commenced on high dose I.V. antibiotics.

Subsequent culture of the tracheal secretions showed gram negative staining in all cases, with staph. aureus isolated in 2 patients.

The 3 intubated patients were extubated after 48 hours. One case, however, required re-intubation for persistent obstruction. All patients recovered and were discharged from hospital within 10 days.

Controversy surrounds the existence of bacterial tracheitis as an independent disease process. Clinically it may be confused with croup and epiglottis. The authors believe, however, that bacterial tracheitis must be considered as a separate entity which requires prompt and accurate diagnosis. It is best managed by early endotracheal intubation with repeated tracheal toilet and aggressive I.V. antibiotic therapy, with close monitoring.

THYROID AGENESIS ASSOCIATED WITH SEVERE GAIT DISTURBANCE

Ann Bergin and Mary King. Temple Street Hospital.

A 14 year old girl presented with a one year history of unsteadiness and progressive deformity of both feet. There was difficulty climbing stairs, running and rising from the kneeling position from early childhood. At 9 years she had painful legs and exercise intolerance, Perthe's disease was diagnosed on x-ray of both hips.

The physical findings were short stature (100 cms), large head (60.6 cms), hypertelorism, kyphosis, cavo varus deformity of both feet, positive Gowers, inability to stand on one leg or tandem walk, peroneal muscle wasting, tight tendo-achilles, weakness of plantar flexion and foot eversion, brisk lower limb reflexes and normal intellect. There was no sensory loss. Investigations revealed normal lower limb EMG and sensory conduction, mildly delayed motor nerve conduction velocity in the lacteral popliteal nerve (41 metres per second), normal myelogram and CT brain scan. The presence of short stature led to consideration of hypo thyroidism with epiphyseal dysgenesis rather than Perthes' disease as a cause of the symptom at 9 years. This was confirmed biochemically and radiologically. Technetium scan showed no uptake in the thyroid region and ultrasound of the neck failed to identify any thyroid tissue. There has been considerable improvement on Thyroxine. This association has not been described previously. The mechanism involved in the neuromuscular symptoms will be discussed.

HYDROHAEMATOMETROCOLPOS – THREE DISPARATE PRESENTATIONS

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Obstruction of the female genital tract may present in a number of ways. We report three cases, one presenting with irritablity and diarrhoea at four weeks, one with recurrent abdominal pain at 12 years, and the third with abdominal distension and backache at 16 years. The anatomy of this condition and associated anomalies will be described.